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CREST SYNDROME

S. PAULINE SHEELA PRIYA M.SC NURSING DEPARTMENT OF MEDICAL SURGICAL NURSING SREE BALAJI COLLEGE OF NURSING, BIHER UNIVERSITY, CHENNAI, TAMILNADU , INDIA.

ABSTRACT:

CREST *syndrome also known as the limited cutaneous form of systemic sclerosis (lcSSc) is a multisystem connective tissue disorder. CREST is a form of Systemic Sclerosis (scleroderma) which is characterized by* ***C****alcinosis (calcium deposits), usually in the fingers;* ***R****eynaud's loss of muscle control of the* ***E****sophagus, which can cause difficulty in swallowing;* ***S****clerodactyly, a tapering deformity of the bones of the fingers and* ***T****elangiectasia, small red spots on the skin of the fingers, face, or inside of the mouth. Quality of life is affected especially with gastrointestinal dysfunction that may cause long term weight loss and malnutrition.*

KEY WORDS*: calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, elangiectasias.*

DEFINITION:

CREST stands for the following:

The acronym stands for Calcinosis cutis, Raynaud phenomenon, Esophageal dysfunction, Sclerodactyly, and Telangiectasias, which are the hallmark features of the disorder.

CAUSES

Limited scleroderma is believed to be an autoimmune disorder — one in which your immune system turns against your own body. In limited scleroderma, the immune system appears to stimulate the production of too much collagen, a key component of connective tissue. This overproduction of collagen builds up in the skin and internal organs, impairing their function.

RISK FACTORS

- **Sex** Women are far more likely to develop limited scleroderma than men are.
- **Race** In the United States, limited scleroderma affects blacks more often than whites. Choctaw Native Americans also have higher rates of limited scleroderma than other races.
- **Genetic factors** If someone in your family has an autoimmune disease — such as lupus, rheumatoid arthritis or Hashimoto's disease — you have an increased risk of developing limited scleroderma.
- **Exposure to toxins.** Certain toxic substances — such as polyvinyl chloride, benzene, silica and trichloroethylene — may trigger scleroderma in people with a genetic predisposition to the disease.

PATHOPHYSIOLOGY

The main pathophysiology of CREST syndrome is the accumulation of collagen and fibrosis in limited areas of the body causing dysfunction. Small arteries in the digits will accrue heavy deposits of collagen, fibronectin, glycosaminoglycan and proteoglycan in the interstitium and intima layer of the blood vessel

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Skin fibrosis is systemically uninhibited causing localized scarring of tissues. Mononuclear infiltrates and CD4 lymphocytes abounds vessel undergoing severe fibrosis connoting an earlier immune response before the onset of fibrosis. The role of collagen stimulator, Transforming Growth Factor – Beta (TGF-B) has been implicated to initiate the symptomatology of calcinosis in CREST Syndrome .

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Pericytes (vascular mural cells) produce excess matrix materials and trigger cytokine activities in blood vessels has been identified to architect the vascular pathologies found in scleroderma . The autoimmune or the “against self” response explains all the pathogenesis that occurs in CREST Syndrome.

Most patients with CREST syndrome have autoantibodies that react with chromosomal centromeres and lack antibodies that react with Scl-70 (topoisomerase I)

SYMPTOMS

While some varieties of scleroderma occur rapidly, signs and symptoms of limited scleroderma usually develop gradually. They include:

Tight, hardened skin In limited scleroderma, skin changes typically affect only the lower arms and legs, including fingers and toes, and sometimes the face and neck. Skin can look shiny from being pulled taut over underlying bone. It may become difficult to bend your fingers or to open your mouth.

On palpation, these lesions are movable and resemble pebbles, ranging from 0.5 to 2.0 cm, underneath the skin.

- **Raynaud's phenomenon** This condition occurs when small blood vessels in your fingers and toes go into spasms in response to cold or emotional stress, blocking the flow of blood. In most people, the skin turns white before becoming blue, cold and numb. When circulation improves, the skin usually reddens and may throb or tingle. Raynaud's phenomenon is often one of the earliest signs of limited scleroderma, but many people have only Raynaud's and never develop scleroderma.
- **Red spots or lines on skin** These small red spots or lines (telangiectasias) are caused by the swelling of tiny blood vessels near the skin's surface. They are not painful and occur primarily on the hands and face.
- **Bumps under the skin** Limited scleroderma may cause tiny calcium deposits (calcinosis) to develop under your skin, mainly on your elbows, knees and fingers.
- **Swallowing difficulties** Poor functioning of the muscles in the upper and lower esophagus can make swallowing difficult and allow stomach acids to back up into the esophagus, leading to heartburn, inflammation and scarring of esophageal tissues.

TESTS AND DIAGNOSIS

- **Blood test**.
- **Barium swallow**
- **Skin biopsy** Sometimes doctors take a small sample of skin that's then examined under a microscope in a laboratory. Although biopsies can be helpful, they can't definitively diagnose limited scleroderma.

MANAGEMENT

Several types of medications can help ease the signs and symptoms of limited scleroderma, including:

- **Topical antibiotics** : If skin ulcers become infected, need to apply topical antibiotics and bandage the area.
- **Antacid drugs** If limited scleroderma is giving heartburn, drugs that reduce the production of stomach acid.
- **Blood pressure lowering drugs** Medications that open small blood vessels and increase circulation may help relieve Raynaud's symptoms and reduce increased pressure in the arteries between the heart and lungs.
- **Drugs to suppress the immune system** These types of medications have shown promise in preventing a condition in which excess collagen collects in the tissue between the lungs' air sacs.

THERAPY

Stiff, painful joints and skin are common problems in limited scleroderma. Learning the right way to stretch and exercise through physical or occupational therapy can help you maintain your flexibility and strength.

- **Physical therapy** Stretching exercises are important to help prevent loss of mobility in your finger joints. A physical therapist can also show you facial exercises that may help keep your face and mouth flexible as well.
- **Occupational therapy** If limited scleroderma is making it difficult for you to perform daily tasks, an occupational therapist can help you learn new ways of doing things.

SURGERY

For some problems, a surgical procedure may be necessary, including for:

- **Calcium deposits** Large or painful calcium deposits are sometimes surgically removed.
- **Red spots or lines** Laser surgery can reduce the appearance of red spots or lines caused by swollen blood vessels near the surface of the skin.
- **Gangrene in fingers** Amputation of fingertips may be necessary if skin ulcers progress to gangrene.

COMPLICATIONS

The chronic fibrosis in CREST Syndrome may cause a perennial drying of mouth, throat and eyes in patients. Dental problems occur with gingival thickening and the acid reflux seen in esophageal dysfunction directly damages the enamel of the teeth. The esophageal involvement can cause frequent acid reflux symptoms and intestinal disturbances like constipation and diarrhea. Malnutrition may ensue with the chronic intestinal fibrotic changes. Systemic fibrosis may affect the heart and cause dysrhythmia. Imaging studies in

CREST patients reveal ecstatic coronary vessels that adversely affects the heart's hemodynamics .

Pulmonary vessels become less efficient with vascular fibrosis leading to lung failure. Raynaud phenomenon in the fingers and toes can cause ulcers and gangrene due to the disruption of the blood flow towards them.

LIFESTYLE AND HOME REMEDIES

Keep warm

To reduce Raynaud's symptoms, wear gloves or mittens outdoors when the weather is cool and indoors when reach into the freezer. To maintain body's core temperature when it's cool, dress in layers and wear a hat or scarf, thermal socks, and well-fitting boots or shoes that don't cut off circulation.

Don't smoke

The best way is to quit Smoke. Nicotine constricts your blood vessels, making Raynaud's phenomenon worse. Smoking also worsens heartburn.

Exercise regularly

Regular exercise can help you maintain your flexibility and strength. Ask your doctor or physical or occupational therapist what activities are right for you.

Change Eating Habits

If patient have difficulty in swallowing, choose soft, moist foods and chew them well. To minimize acid reflux:

- Eat small, frequent meals
- Avoid spicy or fatty foods, chocolate, caffeine, and alcohol
- Don't exercise immediately before or after eating
- Elevate the head of your bed
- Sit upright for two or three hours after a meal

Protection of Skin

Excess collagen destroys sweat and oil glands, leaving your skin stiff and dry. To help soften your skin:

- Avoid harsh soaps and detergents**.** Choose cleansing creams or gentle skin cleansers and bath or shower gels with added moisturizers. Use rubber gloves when doing the dishes or cleaning.
- Reduce bathing frequency**.** Bathe just once a day or every other day, and take brief baths and showers, using warm rather than hot water.
- Moisturize**.** Apply a rich oil-based moisturizer immediately after washing your hands or bathing, while your skin is still damp.
- Apply sunscreen. To prevent further damage to your skin, apply sunscreen before you go outside.
- Use a humidifier**.** Increase moisture levels in home by using a humidifier.

Practice Good Oral Hygiene

- have regular checkups
- use any special rinses or tooth pastes dentist recommends.
- If mouth is chronically dry, try drinking more water and sucking on ice chips or hard, sugarless candy.

Prognosis

The constellation of symptoms from early to late stage is still compatible with life. Disfiguring changes may be delayed with pharmacologic agents if diagnosed early.

Patients with pulmonary hypertension carry a poor prognosis due to its relatively high mortality rate . Quality of life is affected especially with gastrointestinal dysfunction that may cause long term weight loss and malnutrition.

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